Craniofacial Fibrous Dysplasia Addressed Through the Intraoral and Nasal Access

Ivo Cavalcante Pita-Neto¹, Júlio Leite Araujo Junior², Milana Drumont RamosSantana¹, Hermes Melo Teixera Batista¹, Eduardo Costa Studart Soares³, Luiz Carlos De Abreu¹, Luciano Miller Reis Rodrigues¹, Woneska Rodrigues Pinheiro¹, Glauco Soares de Almeida⁴

Abstract

Background: The Craniofacial Fibrous Dysplasia is a benign disease, pseudoneoplastic, rare and asymptomatic of slow growth promoting replacement of bone tissue by an amorphous connective tissue, promoting bone deformation that can invade and cause compression of important structures such as the skull base and facial orbital bone. This study aims to present a clinical case of a Craniofacial Fibrous Dysplasia, emphasizing an alternative of an aesthetic surgical access by an intraoral approach.

Case Report: We report the clinical case of patient IMS, 21 years of age with craniofacial fibrous dysplasia presenting deformity of the left facial middle third in orbital, nasal and maxilla region who was submitted to surgery for facial recontouring. The surgical access was intraoral associated with access for rhinoplasty. A lesion scrape was realized for facial recontouring and nasal osteotomy for correction of deviation caused by the injury.

Conclusions: It was concluded that the surgical technique associated with the intraoral access with nasal extension is an important tool in addressing these lesions, allowing a large surgical field and satisfactory aesthetic and functional results without facial scars.

Introduction

The fibrous dysplasia of jaws has been known for years as a bone injury of the primary hyperparathyroidism, believed by some researchers in 1891 [1]. Only in 1938, after extensive studies observed an isolated
form of the injury, which led them to determine a new terminology, fibrous dysplasia of the jaws, as it is known nowadays [2, 3].

The fibrous dysplasia is a bone neoclassical pseudodisorder, which has affinity for young women, having as a disadvantage its slow asymptomatic and recurrent growth, however it is benign [4]. Depending on their location, it can cause compression of important structures such as the facial orbital bone and skull base. The craniofacial fibrous dysplasia has an amorphous connective tissue caused by the proliferation of fibroblasts interspersed in trabeculae of an immature bone without surrounded osteoblasts. As the spinal bone cavity is gradually replaced by bone fibrous connective tissue in an interlaced form, where spindle cells led to bone deformation. The term fibrous dysplasia of jaws is suitable for neurocranium injuries [5].

The present study aims to present a case of Craniofacial Fibrous Dysplasia, emphasizing an alternative of aesthetic surgical access by an intraoral approach avoiding facial scars.

**Case Report**

Patient IMS, female, without systemic changes, reported swelling in the left cheek when she was 8 years old. It was diagnosed as craniofacial fibrous dysplasia; she was submitted to a facial recontouring surgery. At 17 years old she returns with an aesthetic complaint caused by a further increase in size representing recurrence of the lesion, causing deformation in orbital, nasal and left maxilla region. There was a clear facial asymmetry with nasal, maxilla and dental average line deviation (figure 1). A computed tomography (CT) revealed a radiopaque lesion with a ground-glass appearance in the left jaw region of approximately 7cm of diameter and extension to the base of the skull (figure 2). The neurosurgical evaluation ruled out compression of brain structures. The scintigraphic study showed homogeneous distribution of the radiopharmaceutical in the skeleton, except for the increase of diffuse heterogeneous uptake involving the majority of the left facial bones with increased osteogenic activity. There was no focus of evidence of intense activity of the neurocranium suggesting absence of lesion growth in the prime areas.

A surgical treatment was indicated for removal of the majority of the lesion located in the middle third of the face and to promote facial recontouring. Under general anesthesia an intraoral approach was realized in anterior maxillary alveolar mucosa, nasal mucosa incision isolating the nostrils and union of these incisions with the

![Figure 1: Facial asymmetry with nasal, maxilla and dental average line deviation.](image-url)
intraoral access, allowing the mucoperiosteal detachment in the right and left maxillae, exposing the bone-cartilaginous structures in the nose with its skin shifted upward along the upper lip. The union in the intraoral access with the access for rhinoplasty offered a wide surgical field around the facial middle third and degloving (figure 3a, 3b). Through previous planning by facial prototyping

**Figure 2:** Deformation in orbital, nasal and left maxilla region.

![Figure 2: Deformation in orbital, nasal and left maxilla region.](image)

**Figure 3a:** The Union In The Intraoral Access With The Access For Rhinoplasty.

![Figure 3a: The Union In The Intraoral Access With The Access For Rhinoplasty.](image)

**Figure 3b:** Facial degloving and lesion exposure.

![Figure 3b: Facial degloving and lesion exposure.](image)
(figure 4), there was the removal and scrape off facial external injury promoting facial recontouring, associating rhinoplasty with nasal osteotomies and correction of the deviation caused by the expansion of the lesion (figure 5).

In postoperative follow-up of up to two years, there was a satisfactory cosmetic accommodation of the tissue, improved symmetry and absences of facial scars, making the access of choice an important tool in addressing these lesions with these characteristics (figure 6).

Figure 4: Previous planning by facial prototyping.

Figure 5: Recontouring Associated The Rhinoplasty With Nasal Osteotomies.

Figure 6: Satisfactory cosmetic accommodation of the tissue, improved symmetry and absences of facial scars.
Discussion
Several studies have been devoted to the study of fibrous dysplasia of the jaws, but the study in question shows a rare condition of simultaneous involvement in cranial and facial region [6, 7].

The craniofacial region is an important site for the pathology onset, being the maxilla and mandible the featured sites. The patient had no evidence of lesion in the mandible [8].

The fibrous dysplasia of maxillae can be divided into two groups, monostotic and polyostotic. The monostotic form is considered the most common, affecting only one bone or contiguous bones, corresponding to approximately 70% of cases [8, 9]. When the disease occurs in two or more bones without continuity, it corresponds to the polyostotic form, representing 30% of cases. When the fibrous dysplasia appears accompanied by skin pigmentation of the type of "coffee with milk" and endocrine diseases it is called McCune-Albright syndrome [10].

In patients with fibrous dysplasia presenting the polyostotic form, it was observed that most of these patients presented craniofacial involvement, being one side of the body most affected. In a better way, most patients with fibrous dysplasia in the craniofacial region have the monostotic fibrous dysplasia form as prevalent [9].

The observations made by other authors are consistent with relation to the appropriate designation for neurocranium involvement [5, 11].

With regards to the standard states that the monostotic form has no predilection for gender. However the polyostotic form has a predilection for females [8].

Regarding the etiology, the literature is also confusing and divergent. The author Lichtenstein [2], believes there is an abnormal enzymatic activity in the process of mesenchymal bone formation. But Murray [12], supports the hypothesis that there is a change in calcium and phosphorus metabolism. Attention to the occurrence of osteoblastic hyperplasia [13]. However the monostotic form of this disorder could not be correlated with the syndromes of Albright and polyostotic fibrous dysplasia [14].

There are several divergences related to its pathophysiological, where several authors defend the idea that the monostotic fibrous dysplasia is a deviation from the normality of the mesenchymal bone formation process, being a congenital abnormality of dominant or recessive autosomal transmission [14].

Malignant and benign lesions make themselves as differential on the diagnosis of craniofacial dysplasia of the jaws. Injuries such as, solitary unilocular cysts, not ossifying fibroma, eosinophilic granuloma, cholesteatoma, meningioma, Paget's disease, osteochondroma, ossifying fibroma, giant cell reparative granuloma, exostosis, aneurysmal bone cyst, cystic fibrous osteitis, ameloblastoma, and plasma cell myeloma. Injuries such as sarcoma and metastatic osteoblastic lesions are considered malignant lesions of diagnosis. Observed an occurrence of 0.4% of spontaneous neoplastic transformation to sarcoma [15, 16].

The craniofacial fibrous dysplasia can be treated by surgical technique and / or drugs. Being the surgery indicated only for symptomatic cases or in cases of functional changes, anatomical distortion of the bones involved, to prevent bone expansion and pathological fractures [17, 18].

The case of fibrous dysplasia, the radiotherapy becomes a contraindication because it can increase the malignant degeneration, not being this a predictor factor of malignancy, since in rare cases, the spontaneously sarcomatous transformation has been reported. Radiotherapy was not contemplated to the patient, and the emphatic literature contraindicates its use. The patient is being evaluated periodically because of the possibility of recurrence and she has been oriented [3, 19, 20].
Conclusions
The fibrous dysplasia is a lesion that has several characteristics in common with other fibro-osseous pathologies. It is of extreme importance to get as much information, through clinical, imaging, laboratory and histopathological examinations to reach a correct, early diagnosis and appropriate treatment. Several discrepancies was observed in the literature and it was concluded that the Craniofacial Fibrous Dysplasia is a fibro-osseous pathology that still has several obscure fields because of lack of research.

For the dental surgeon, it is important to have knowledge of this condition, as this disease can affect orofacial structures.

Ideally, the treatment should be performed after the growth period is finished, however, in cases involving the aesthetic and functional aspects severely; it is advisable to intervene before adulthood. The follow up is very important, being important in the early detection of possible recurrence, reactivation or malignant changes.

Consent
Written informed consent was obtained from the patient for publication of this Case report and any accompanying images.

Ethics questions
The ethical committee of the Regional Hospital of Cariri, for case presentation, approved the current study.

Competing interests
The authors declare no conflicts of interest. All research was conducted with their own resources.

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References


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